▪ Headache Medicine 2020, 11(2):32-34 ▪ ISSN 2178-7468, e-ISSN 2763-6178

Headache Medicine

▪ DOI: 10.48208/HeadacheMed.2020.10

Clinical Correspondence

Vessel wall imaging for diagnosis and follow-up of basilar artery reversible cerebral vasoconstriction syndrome (RCVS)

Matheus Kahakura Franco Pedro **Paulo Sérgio Faro Santos** **Bruno Augusto Telles**

Department of Interventional Neuroradiology, Instituto de Neurologia de Curitiba, Curitiba, Paraná, Brazil.

Matheus Kahakura Franco Pedro
matheuskfpedro@hotmail.com

Edited by
Marcelo Moraes Valença
Mario Fernando Prieto Peres

Keywords:
Vasoconstriction
Magnetic Resonance Imaging
Vascular Headaches

Abstract

Reversible Cerebral Vasoconstriction Syndrome (RCVS) is a clinical and radiological syndrome that is primarily defined by thunderclap headache, with or without further neurological deficits, and segmental intracranial vasoconstriction that resolves within three months. The current nomenclature was only established in 2007, but it has been known with diferent names for over fifty years. The pathophysiology, while still not completely understood, seems to point towards a disease based on abnormalities of vascular tonus without structural inflammation. It is clear, however, that patients with RCVS often have triggers, especially drugs or other vasoactive substances. Distinguishing this entity from others, especially subarachnoid hemorrhage and arterial dissection, is extremely important, given the particular prognosis and need of immediate treatment of each disease. The preferred imaging method has long been the angiography; however, new magnetic resonance imaging (MRI) such as vessel wall imaging have allowed for non-invasive diagnosis and follow-up. The authors report a case in which MRI was used in a patient with basilar artery RCVS and present a literature review.

Received: April 2, 2020.
Accepted: April 27, 2020.

Introduction

Reversible Cerebral Vasoconstriction Syndrome (RCVS) is a clinical and radiological nosologic entity that is primarily defined by hyperacute onset of thunderclap headache and segmental intracranial vasoconstriction that resolves within three months, with or without further neurological deficits[1]. Though digital subtraction angiography has long been the standard work-up exam, the role of magnetic resonance imaging, particularly after the refinement of vessel wall imaging, has substantially expanded[2,3].

Case Report

The authors present the case of a 31 years-old Caucasian female with no previous history of headache, who presented to the ER due to a sudden, thunderclap occipital headache while performing strenuous physical activity (cross-fit). No other neurological symptoms or deficits took place. She underwent an arterial angiotomography which suggested vascular lumen reduction of the basilar artery. Her laboratory work-up showed no noteworthy alteration. Afterwards, she underwent brain MRI with vessel wall imaging on a 3-Tesla machine, which confirmed a stenosis inferior to 50% on the middle section of the basilar artery along with gadolinium enhancement towards the vertebro-basilar junction (Figure 1). Her headache receded without need for medication and she was released for outpatient follow-up. After three months without any symptom, another brain MRI with vessel wall imaging on the same machine was performed, showing near complete resolution of the stenosis, as well no further enhancement on the basilar artery after gadolinium injection (Figure 2). On the same outpatient visit, the patient reported regular use of a performance enhancement compound including caffeine and bupropion.

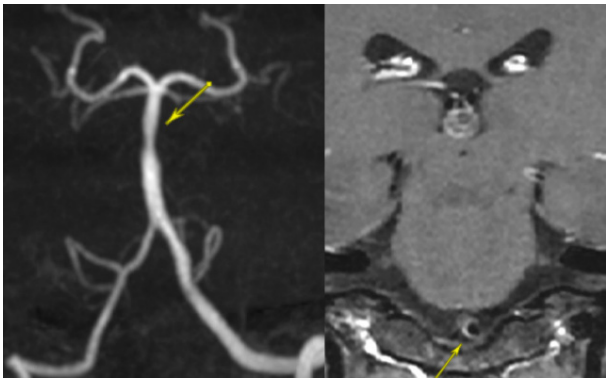

Figure 1. Left - coronal view of reconstruction of arterial angioMRI on a 3 Tesla magnet confirming stenosis of the middle third of the basilar artery (arrow); right - coronal slice of vessel wall imaging after gadolinium injection, with impregnation of the basilar artery near the vertebro-basilar junction (arrow).

showing complete resolution of the gadolinium enhancement by the vertebro-basilar junction.

Discussion

Though this nosologic entity was first reported over fifty years ago, its most consistent description came in 1988 by Call and Fleming[4]; Calabrese proposed the current nomenclature in 2007 and established formal diagnostic criteria, thus unifying the many "diseases" with similar clinical and radiologic features under a single term.[5] No precise data on incidence is currently available, though it doesn't appear to be particularly rare[6]. The pathophysiology remains a mystery, although alteration on vascular tone leading to vasoconstriction seems to be the main mechanism[1], which is supported by the lack of vascular or perivascular histological abnormalities on biopsy of brain tissue. The role of sympathomimetic vasoactive substances is well known, with caffeine and bupropion having been previously recognized as triggers[7,8]. The differential diagnosis includes subarachnoid hemorrhage, cervical arterial dissection, and primary angiitis of the central nervous system; as such, correct differentiation between these entities is of paramount importance, given the different mechanisms and treatments. In terms of prognosis, the disease is monophasic and typically self-limiting, with the criteria establishing resolution within three months.

Conclusion

This case illustrates the typical course of the disease and the need to recognize it and differentiate from other vascular diseases of the central nervous system. The use of MRI with vessel wall imaging allows for both accurate diagnosis and follow-up in a non-invasive manner.

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